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Retroperitoneal extra-adrenal ganglioneuroma involving the infrahepatic inferior vena cava, celiac axis and superior mesenteric artery: A case report

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ABSTRACT

INTRODUCTION: Ganglioneuromas are rare benign neoplasms arising from the sympathetic neuroendocrine system. These tumors usually occur in the abdomen and tend to grow around major blood vessels making often their complete excision challenging and demanding.
PRESENTATION OF CASE: The authors present the challenging surgical management of a sizable retroperitoneal extra-adrenal ganglioneuroma involving the infrahepatic inferior vena cava, portal triad, celiac axis and superior mesenteric artery in a 23-year-old female patient. The tumor was safely and completely excised in toto with preservation of all neighboring vital anatomical structures using a midi laparotomy access.
DISCUSSION: Ganglioneuromas should be included in the differential diagnosis of any retroperitoneal mass. Their management involves total surgical excision however, in some instances; it can be challenging and demanding because of their tendency to engage neighboring vital anatomical structures.
CONCLUSION: A surgical strategy including meticulous operative dissection guided by the quality principles of surgical oncology although challenging and demanding can result to a safe and complete tumor excision, which is directly correlated with an improved patients' postoperative outcome and excellent prognosis.

1. Introduction

Ganglioneuromas are rare benign neoplasms arising from the sympathetic neuroendocrine system. These tumors usually occur in the abdomen and tend to grow around major blood vessels making often their complete excision challenging and demanding.[1–3]

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the superior mesenteric artery in a 23-year-old female patient. The tumor was safely and completely excised in toto with preservation of all neighboring vital anatomical structures using a midi laparotomy access.

2. Presentation of case

A 23-year-old female presented initially to her family physician complaining for intermittent abdominal epigastric discomfort and pain of 6-months duration. The patient had no weight change, hypertension, or genitourinary symptoms. Her family history was negative for cancer or other neoplasias. She did not have any previous surgical interventions. Physical examination was normal, without hypertension or palpable mass in the abdomen. An ultrasound revealed a solid mass located in the upper most portion of the omental bursa. Computed tomography (CT) scan depicted a retroperitoneal mass measuring 6.3 cm × 3.8 cm × 4.5 cm that was compressing/involving the infrahepatic inferior vena cava (IVC).

Laboratory workup performed at our hospital included normal blood count, serum electrolytes and serum cortisol. Endocrinologic evaluation combined with 24-h urinary levels of vanillyl mandelic acid (VMA) and metanephrines were normal. A CT-guided biopsy of the mass revealed a fibrous and mixed neural tissue of benign histology consistent with ganglioneuroma. After obtaining the patient's consent, she was elected to have a surgical resection.

At surgery, the abdomen was opened through a "midi" (<8 cm) upper midline incision and the round and falciform ligaments were divided. The left hemiliver was mobilized by dividing the left triangular ligament. This was followed by the division of the inferior leaf of the hepatogastric ligament up to the level of ligamentum venosum arantii, in order to expose fully segment I of the liver and infrahepatic IVC (Fig. 2). No aberrant left hepatic artery was present and special attention was given to preserve the hepatic branch of the anterior vagus nerve.

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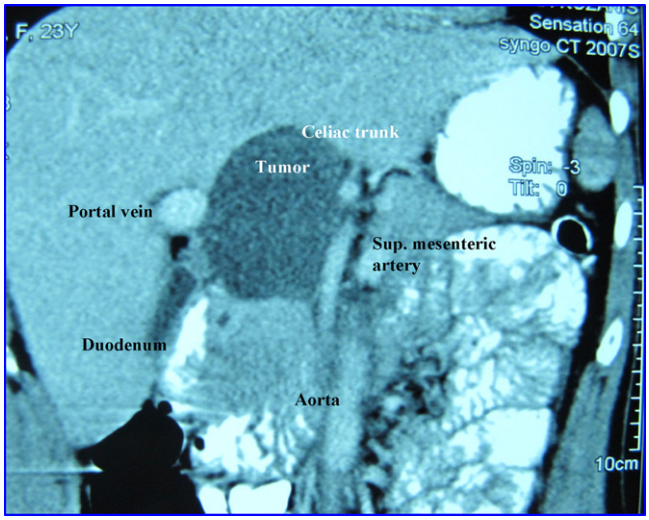

Fig. 1. Computed tomography scan depicting a retroperitoneal mass measuring 6.3 cm × 3.8 cm × 4.5 cm compressing/involving the infrahepatic inferior vena cava, portal triad, celiac axis and superior mesenteric artery.

These steps exposed the upper margin of an oval shaped, yellowish, firm, mass, which was arising from the retroperitoneum adhering to the caudate lobe and infrahepatic IVC. Following this, the compressed by the tumor hepatoduodenal ligament was encircled by a penrose drain and retracted to the right of the midline to facilitate the mobilization of the tumor's right margin away from the structures of the portal triad. After meticulous preparation and division of loose adhesions, the plane of cleavage was created between the tumor's pseudocapsule and adjacent structures, and the tumor was safely mobilized away from caudate lobe, IVC, portal triad, and left gastric artery. Caudally, the tumor was draping the aorta involving the celiac axis and the origin of the superior mesenteric artery. While maintaining the dissection in the cleavage plain, preserving as far as possible all neighboring nerve tissue the tumor was safely and completely excised (Figs. 2 and 3). The patient had an uneventful postoperative course and was discharged 8 days after the surgery. As part of her annual follow-up examination she was scheduled to undergo an abdominal MRI screening.

Tumor's histopathology revealed that it consisted of spindled cells, with indistinct cell borders in a fibrillar matrix, which

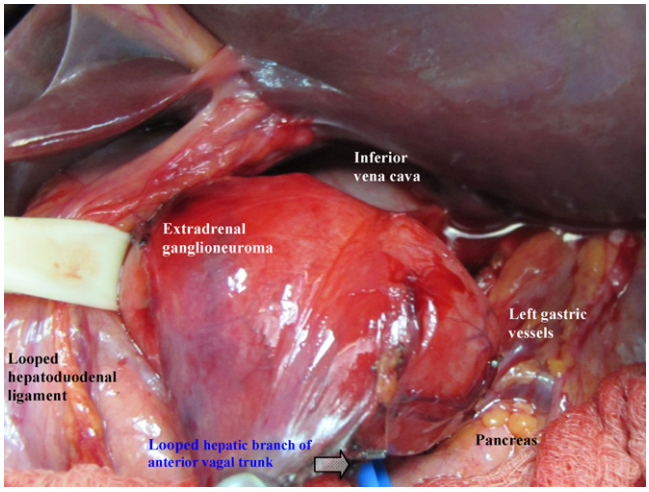

Fig. 2. Intraoperative image: mobilization of the upper margin of an oval shaped, yellowish, firm tumor arising from retroperitoneum adhering to the caudate lobe and infrahepatic IVC.

Fig. 3. Cut surface of the surgical specimen. The mass had a white cut surface with a focally edematous appearance, measuring 7 cm × 4 cm × 3 cm.

contained varying numbers of ganglion cells (Fig. 4a). Ganglion cells had large round nuclei with prominent nucleoli and abundant eosinophilic cytoplasm. No atypia or mitotic activity was evident. The ganglion cells stained positive for S-100 protein while Ki-67 was <1% (Fig. 4b). On the basis of these histopathological findings, the final diagnosis of an extra-adrenal ganglioneuroma was established.

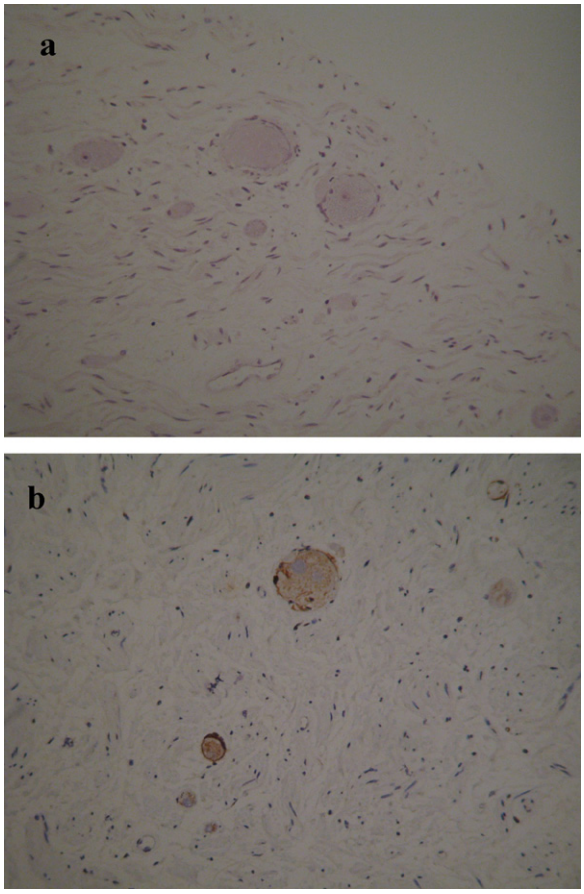

Fig. 4. (a) Microscopic scrutiny of the resected specimen. Multiple ganglion cells in a dense matrix of Schwann cells (H&E; ×400). (b) Ganglion cells stained positive for S-100 protein while Ki-67 was <1%.

3. Discussion

Ganglioneuromas are usually benign differentiated tumors of the sympathetic nervous system. They originate from embryonic undifferentiated cells of the neural crest and are part of the wider spectrum of neuroblastic tumors, which also includes neuroblastoma and ganglioneuroblastoma.[1] These tumors can grow at any site of the sympathetic nervous tissue but they usually occur in the abdomen. Fifty-two percent of ganglioneuromas are located in paraspinal retroperitoneum (sympathetic ganglia), 39% in posterior mediastinum, and 9% are located in the pelvis or neck.[2,3] Of the abdominal ganglioneuromas, 49% originate in the adrenal gland and 51% are extra-adrenal.[3] In some cases, ganglioneuromas are the final stage of maturation of neoplasms such as neuroblastoma or ganglioneuroblastoma. Nevertheless, many of these tumors appear to arise de novo based on age at diagnosis (over 10 years of age), and anatomic location.[3]

The relatively low incidence of ganglioneuromas often makes their diagnosis elusive. Usually, they are clinically silent, regardless of their size and should be included in the differential diagnosis of any retroperitoneal mass. Similarly, clinical signs and symptoms of hormone excess are usually absent, although ganglioneuromas are hormonally active producing a wide range of neuropeptides. Patients may rarely present with vague abdominal pain or neurological symptoms due to mass effect.[1–3]

Ganglioneuromas are typically discovered during routine imaging studies. On computed tomography scan[4] they appear well delineated and they tend to grow around major blood vessels without narrowing them. On magnetic resonance, ganglioneuromas appear homogeneous and have relatively low signal intensity on T1-weighted images. On T2-weighted images, the signal intensity is proportional to the ratio of myxoid stroma to cellularity as well as to the amount of collagen present at the tumor site.[5] Although, iodine-tagged metaiodobenzylguanidine uptake can be increased in patients with ganglioneuromas it does not discriminate the exact type of the catecholamine-producing tumor. Diagnosis can be established after a CT-guided biopsy,[3] as happened in the present case.

Their management involves total surgical excision when feasible however, in some instances; it can be challenging and demanding because of their tendency to engage neighboring vital anatomical structures. The prognosis is excellent for ganglioneuromas after surgical removal.[2,3,6,7] After being completely excised, they do not usually recur, although malignant transformation into peripheral nerve sheath has been described.[6] After tumor excision, immunoperoxidase staining test of the tumor ganglion cell can also add to diagnosis, showing a strong S-100 protein positivity for neuron-specific enolase and spindle-cell component.[3] The slow-growing pattern of the tumor necessitates long term follow-up of operated patients, including repeated imaging along with careful physical examination for local recurrence or new symptomatology.

In conclusion, a surgical strategy including meticulous operative dissection guided by the quality principles of surgical oncology although challenging and demanding can result to a safe and complete tumor excision, which is directly correlated with an improved patients' postoperative outcome and excellent prognosis.

Conflict of interest statement

None.

Funding

None.

Ethical approval

The patient has been informed and we state that we have obtained written consent from the patient and that we can provide this should the Editor ask to see it.

Author's contributions

K. Vasiliadis has contributed for the study design, data collections, data analysis, and writing. C. Papavasiliou, D. Fachiridis, and M. Kiranou have collected data for the study. S. Pervana and M. Michaelides have analysed the data and C. Makridis has contributed for the study design.

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